

## **EB AND PAINMANAGEMENT**

### **JEB generalized intermediate, JEB others (rare)**

#### **1. Introduction**

In EB there may be many different reasons for the occurrence of acute and or chronic pain. You should know ahead of your assessment which form of EB the patient has: In our experience, you may experience pain, it is common in EB, but it is not the mandatory rule! With the development of pain it is necessary to look closely at each individual case and what are the reasons for the pain as they may vary in each type of EB. In the best of circumstances you can find ways to reduce or eliminate pain. It all depends on the form of EB and reasons for the discomfort.



#### **Important points in a nutshell**

- **Causes for pain in EB**
  - **The condition itself (blisters, wounds, difficulty swallowing ...)**
  - **The treatments (lancing blisters, dressing changes ...)**
  - **Medical interventions (drawing blood, injections, surgery ...)**
  - **Additional reasons for pain (headaches, stomach aches...)**
- **Therapy:**
  - **You should always treat the underlying cause of the pain first.**
  - **All dressing changes should be performed as pain free as possible.**
  - **When dressing changes have the potential to cause pain, evaluate the need for medication or alternative therapies for treatment including psychological measures. Remember, it is often more effective to combine different therapies.**

## 2. General information

Pain has an important function for our bodies. When we feel pain our body is signaling us that something is not right. When we feel pain our brain shifts our focus immediately to whatever is causing the pain. For example, when we feel acute pain in our hand we quickly look towards our hand to see what the cause of the pain is. When we see that our hand is over a candle with a burning flame we see what is causing the pain and pull our hand quickly away from it.

Acute pain gives us protection through its warning system and it is important for the integrity of our bodies. Our bodies warn us when something is not right and it is important not to ignore these signals and we should investigate where exactly the pain is coming from. Many patients see a doctor for the first time due to pain and are often newly diagnosed with a disease.

In addition, it is also possible that the pain will occur more frequently, will be stronger and last for a longer period of time. Then you slowly lose this warning and protective function. The pain will be chronic, and for many people a tormenting life companion. To have pain continuously and repeatedly, usually implies a major limitation to your quality of life.

Both in acute and chronic pain - it is necessary to look at the pain closely to understand what the causes are, and in the best case find ways to be able to reduce or eliminate it.

In EB the occurrence of acute and chronic pain can be caused from a variety of things. From our experience one thing should be said: no matter which form of EB it is, **you may** experience pain, it is common in EB, but it is **not** the definite rule!

The causes of pain can be variable. On one hand, there is pain caused by the disease itself, such as fresh wounds, but also pressure points that may arise due to large fluid-filled blisters. On the other hand, the treatment of the consequences of the disease can be painful. Examples would be the opening of blisters, peeling off adhesive bandages or from required surgery. In addition, all people living with EB, can also suffer from other types of pain. Often these have nothing to do with their underlying disease, but are also very uncomfortable, such as migraines, back pain, dental pain, abdominal pain, etc.

With a chronic illness such as EB, minor and major medical procedures often need to be performed on a regular basis. Blood tests are repeatedly performed or a medication needs to be given intravenously or by injection. Many tests are necessary as in biopsies (skin samples) and various operations. Proper pain management must be planned for every procedure.

One specific issue related to all types of EB is the pain that occurs due to the treatment. When dressings adhere to wounds, when wounds are infected, or when disinfecting ointment must be applied, this is painful. Not just occasionally, but regularly, several times a week, throughout their lives.

There are additional challenges: the oral hygiene is often difficult, regular physical exercise should be routine yet it can also cause pain. This does not affect all forms of EB to the same extent and also changes repeatedly in the course of life, but the majority of EB sufferers and their families deal with this issue again and again on a regular basis.

Pain in newborns and infants with EB are a separate issue and are dealt with in a special chapter "neonatal care".

It is known that the sensation of pain can be quite different for every individual. This has many causes, which in individual cases often cannot be accurately determined. But one thing is true for all people with pain: only he or she knows themselves how painful anything is! What one person sees as barely causing a problem can be very distressing for another person. A familiar example is taking blood samples or changing the dressing for people with EB, each person has a different perception of how painful these procedures are. It is important to find a way to help each individual deal with their pain.

There are a number of factors which can increase the pain. With a chronic illness such as EB it is especially important to take care of these factors wherever possible, particularly to prevent unnecessary excessive pain. Fear, anxiety, depression and other unpleasant feelings are the best known of these factors. Each of us knows an existing headache can be aggravated, if you have stress or worries. But the same goes for any other type of pain and is therefore a major problem that is often reinforced from each other. More worries lead to more pain, and then there are more worries about, and so on...

Also the feeling of being at the mercy of pain you cannot control and the uncertainty about what there is right now in front of you may be pain-enhancing. Physical exhaustion due to the disease itself and other symptoms, such as nausea, fatigue or shortness of breath may also play a role. Sometimes, even a low-stimulus, boring or not age-appropriate environment that lacks diversion can enhance the perception of pain. Therefore, pain is more intensively perceived at night.

In EB the issue of pain can be a very complex subject. You should know that there are good solutions for many of the problems that can arise. A good collaboration with experienced physicians, highly trained therapists and nursing staff is very important.

### **JEB generalized intermediate, JEB others (rare forms):**

**In JEB generalized intermediate/JEB others (rare forms) the following causes can repeatedly lead to pain:**

#### **Blisters:**

In EBS generalized intermediate pain can be directly caused by the blisters. Especially large, unopened blisters on the soles of the feet can be extremely unpleasant. The pain is usually due to the pressure exerted by the blisters on the surrounding body structures such as connective tissue, muscle or bone. Blisters may also be painful elsewhere on the body, they usually occur in places where there are pressure points from clothing (e.g. diapers or a belt).

#### **Wounds:**

In addition, open wounds can be painful. If, for example, after lancing a large blister, the blister roof was removed, it may be that the open wound can be quite painful. Such wounds should be covered quickly with an appropriate wound dressing; this will also help relieve the pain. In addition, some individuals report that they feel a slight pain when a wound heals. This may also occur in combination with itching.

#### **Infection:**

Occasionally a wound can become infected and or inflamed. An infection is caused by the fact that germs in the tissue (in this case in the region of a blister or wound) proliferate. These germs are usually bacteria, fungi, viruses and protozoa and can

occur in exceptional circumstances. Warnings for inflammation are mainly **pain**, **heat** (locally at the wound site or generalized as fever), **redness** (around the blister/wound, or red lines), and occasionally yellow or honey-colored scabs and the presence of pus.

#### Pain in the stomach and intestines:

One reason for pain is also problems in the digestive tract. Stomach pain can occur caused by inflammation, severe bloating (e.g. as a side effect of medication or supplement drinks), or pain due to constipation and very hard stools. Anal fissures may occasionally occur (small tears in the anus), which are very uncomfortable and painful.

#### Pain when eating:

With involvement of the mucous membranes injury may also occur in the form of blisters and open sores in the mouth. Biting, chewing and swallowing can thus be associated with pain. This is usually easy to control, and rarely leads to larger problems. It may also to be confused with other diseases, such as the extremely unpleasant and painful aphthae (small mouth ulcers), which many people suffer from. Due to enamel defects, which are often seen in this form of EB, you can sometimes suffer from aggravating tooth pain making eating difficult.

#### Eye pain:

A particularly painful problem are blisters that form around the eyes. In JEB generalized intermediate, this occurs in only a few cases, but it is very unpleasant. In most cases these are small blisters and ulcers on the cornea, the eyelids may also be affected. The reason for the formation of these wounds is unknown, and little can be done to prevent them. Avoiding wind, cold and not rubbing your eyes when tired or sleepy has not prevented formation of these eye blisters. .

#### Bone pain, joint pain, contractures:

In severe forms of EB, due to a combination of reduced mobility, poor posture, muscle weakness, malnutrition and limited exposure to sunlight pain may occur. Genetic factors for osteoporosis and other conditions involving the joints may also be contributing factors. This usually results in pain, both directly in the joints and bones as well as indirectly by contractures of the joints.

In JEB forms these changes are usually not very pronounced, but should be assessed and documented.

#### Other causes:

When the onset of pain occurs for other reasons, with JEB generalized intermediate you can normally assume that the cause is not EB. Of course, people with EB suffer occasionally from flu, migraine and perhaps appendicitis. Unfortunately, all other diseases and illnesses can be contracted regardless of whether you have EB or not. Contact your family doctor or pediatrician who will treat you further.

### 3. Therapy in general

Depending on what form of EB you or your loved one is suffering from, sometimes you'll have to deal with it more intensively. The principles of pain management apply to all forms, just in varying degrees.

Pain that occurs locally, in a limited area, should only be treated locally. Pain which occurs in several areas or affecting the whole body should be treated as a whole.

When the cause is found for the pain various treatment options are available. Ideally you should initially, treat the underlying cause. This means, for example, to avoid pressure points from clothing, or regularly carry out a professional dental cleaning. Even at an early stage the start of regular exercise can have a pain-reducing effect, by improving bone preservation and mobility.

You should think and plan for the long term. Pain therapy in a disease such as EB is not always the most effective. In many cases it may not be possible to fight the cause of pain in fact, we have to resort to a series of other measures for good pain management. To keep this chapter manageable we only have a summary of the general measures, so that successful pain therapy might be possible.

To keep this chapter manageable we only give a summary of the general measures for successful pain therapy that are possible. For specific topics for EB, such as dressing changes, eye pain and pain in the gastrointestinal tract please read the respective chapters that deal specifically with these issues! Also the treatment of pain in the context of medical treatment, such as the insertion of an intravenous line or by surgery, we treat in detail in another section.

## Wound Care:

Independent of the form of EB the most important and decisive action to reduce pain in EB is proper wound care. Dressing changes must be made as painless as possible, which is already an essential step towards pain reduction. It is well known that you cannot use adhesive materials and band-aids for the dressing of wounds for EB cases. The materials that are designated as "non-adhesive" may also not be suitable. In addition the best dressing materials can stick due to dried wound exudates and drying of creams and ointments. When the dressing sticks and has dried onto the wound and needs to be removed it can become painful.

With experience one must note that wounds exposed to room temperature are much more painful, than a dressed wound. Therefore the dressing changes should not take more time than absolutely necessary. The practice for dressing changes is that it needs to happen quickly. In many cases it is better to treat only one area at a time, for example to remove the dressings from an arm and then immediately to apply a new bandage. This followed with dressing the next arm, then the first leg, etc. Many of our patients like to remove all their dressings at once in a warm bath. The dressings then need to be applied very quickly in a warm room after the wounds are well dried.

You should not underestimate the role of the person who performs the dressing changes. A self assured person, who can work quickly, will help to make the routine dressing changes be painless.

You need to find the best method for yourself or your loved one by working together with specialized doctors and nurses trained in wound management. In our view you cannot develop a "standard" to be applied equally to all people. On one hand, the needs are very different (depending on the form of EB, but also by the phase in life and personal preferences). On the other hand - and this is a point not to be underestimated - many materials are not available in all regions and countries of the world. The best material is useless if it is not available and/or not affordable. Alternatives must be considered, which are applicable and realistic. For more information and suggestions, see the section entitled "Wound Management".

## Medications:

One generally thinks first about medications for pain therapy. If all preventative measures have been implemented and still the pain persists, then we have a number of good medications for pain therapy in EB in various strengths. With a genetic disease which currently is not yet curable, such as EB we must also think about the long-term effects and especially the side effects of drugs. If pain medications are necessary, the appropriate medication with the correct dose should be taken for a long enough period of time to be effective. Since it is very different to know which drugs are suitable in what dosage, and for which age group for an effective medical pain therapy and you may encounter side effects, it must always be prescribed by a physician or a health care professional. Unfortunately all good and effective medications have side effects. Since with EB it is often necessary to take medication for a long time, you must pay special attention to the side effects.

With some medications check-ups are necessary at regular intervals. Often a combination of pain medications has to be taken along with other drugs. Combinations of certain medications can be dangerous or cause some medication to become ineffective. We would like to advise you not to take over the counter medications without speaking to your doctor first.

Basically, the medical pain therapy for EB has the same schema that applies to all other pain treatment. A very helpful so-called step scheme (WHO = World Health Organization) is well known by every doctor. It recommends beginning gradually starting with weaker medication for effective pain relief, then a stronger medication and increase it until sufficient pain reduction is achieved. The same scheme can also apply for EB. There is a limitation on the type of preparation of the prescribed medications. Some affected EB patients cannot swallow any pills. In such cases it is recommended that painkillers are prescribed in a liquid form, such as syrups or drops. Occasionally, an analgesic which may be applied as a gel may be used for the treatment of individual areas.

### **There are 3 other medication limitations that should be considered in EB:**

1. Azetylsalicysäure is not suitable for pain therapy in EB. Besides the fact that it should never be prescribed before the onset of puberty, it also leads to an increased



risk of bleeding, so that it can result in considerable loss of blood from the wounds. There can be exceptional cases, but in general we do not recommend the choice!

2. Medicines that contain Acetaminophen, have little to no effect on pain caused by inflammation. Since EB wounds often have an inflammatory component, such drugs usually have little effect on wound pain. However for other types of pain the drug can be used with good effect.

3. For understandable reasons no pain patches should be used with EB. However, there may be exceptional cases.

### Non-pharmacological measures:

The use of medication is not always necessary; there are also a number of other measures that can help to reduce pain. There are many other [physical](#) and [psychological methods](#) that can be effective. Also, methods from the field of complementary medicine have often proven themselves helpful to some affected people. It is important that you seek advice from your physician. They know best what treatment possibilities there are and where they are available, where you can try to learn the different methods and how the different methods can work together. It will often be helpful to seek advice at a special pain center or a pain clinic in your community. Here is a brief overview of the measures that have proven effective for EB in several cases.

### **This is a list of only some of the possibilities that can be effective:**

#### [Psychological measures:](#)

- Distraction in infants by dominating the environment with visual stimuli (bubble blowing toys, musical mobile) and later all kinds of sensory stimuli (storytelling, picture books, view reflected images, ... music, video or television)
- Learn relaxation techniques under special guidance (autogenic training, progressive muscle relaxation according to Jacobson)
- Hypnosis (learning self-hypnosis has been experienced as especially helpful by those affected)
- Parental education programs

- Explanation of everything in a child-and age-appropriate way including everything that is disease related (this must not in any case be underestimated, because the fear of the unknown can significantly increase pain)

#### Physical methods:

- Cold or heat (for example, cooling baths with aching feet, or hot water bottle for abdominal pain)
- Low-energy laser (this can be especially helpful with milder forms of EB to help reduce pain)
- Delicate massages
- Slight vibration
- Swimming – exercise in water

#### Complementary medicine methods:

- With these methods, there is often no proven scientific background, but some people experience a tangible improvement.
- Homeopathy
- Acupuncture/Acupressure
- Biofeedback

To be able to judge the success of a certain therapy for your pain it can be helpful to keep a pain diary. It is best if the person makes an entry daily of where and when the pain occurs, an actual time is helpful. What time the action took place and the drug that was taken should be noted, as well as the severity of the pain. The use of a pain scale makes it possible even for younger children to use a diary. For very young children you still have to make do with someone else's judgment. The assessment of pain intensity by the parents has been shown to be more meaningful than the way the doctors/nursing are judging the child's pain.

We are forced to deal with pain as a painful companion. Our goal is not to be completely free from pain by continuous medication. Whenever possible we will try to get along with non-pharmacological measures, and in many cases we will opt for the combination of several options. The principle is: The more you can avoid drugs, the better. Therefore, prevention and avoidance of pain is the most important approach to good pain management, followed by non-drug measures, and the last option is medication. With good cooperation from all the interested parties and a large dose of patience, a sufficient pain therapy for all forms of EB can be made possible in many cases.